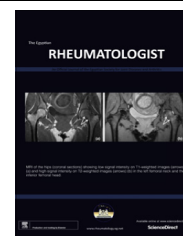




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#### CASE REPORT

# Post Chlamydial reactive arthritis in a case of Vogt-Koyanagi-Harada syndrome (VKH) with negative HLA-B27: An association or just coincidence



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#### KEYWORDS

Vogt-Koyanagi-Harada (VKH);  
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 Ulcerative colitis;  
 Seronegative spondyloarthropathy (SpA)

**Abstract** *Background:* Vogt-Koyanagi-Harada (VKH) syndrome is a multisystem disorder characterized by granulomatous panuveitis with exudative retinal detachments that is often associated with neurologic and cutaneous manifestations.

*Aim of the work:* The aim of this case report is to describe a rare case with Vogt-Koyanagi-Harada syndrome that developed an explosive form of reactive arthritis shortly after attack of Chlamydial urethritis. An association between Vogt-Koyanagi-Harada and ulcerative colitis was previously described in several case reports. The case is described in detail and the literature was reviewed.

*Case report:* In this report we described a male patient with long standing Vogt-Koyanagi-Harada syndrome, who developed aggressive reactive arthritis two weeks after an attack of Chlamydial urethritis. Clinically the patient presented with bilateral sacroiliitis, peripheral arthritis, and wide spread enthesitis. The patient had positive family history of scleroderma in his first degree relative and HLA-B27 testing was negative.

*Conclusion:* In this report we theoretically proposed a possible relationship between VKH and Seronegative spondyloarthropathy group of disorders.

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#### 1. Introduction

Vogt-Koyanagi-Harada (VKH) syndrome is a rare disease of the eyes characterized by bilateral uveitis (inflammation of the uveal tract consisting of the iris, ciliary body, and choroid), alopecia (hair loss), poliosis (depigmented eyelashes, eyebrows, or hair), vitiligo (a skin disease with depigmentation), and hearing loss [1].

Interestingly the Akita breed of dog (animal model of VKH) is affected by a number of immune-mediated diseases, including autoimmune thyroiditis, sebaceous adenitis, pemphigus foliaceus, uveitis, polyarthritis, myasthenia gravis, and uveodermatologic (UV) syndromes, myasthenia gravis, juvenile polyarthritis/meningitis, discoid lupus (0.5%), and systemic lupus erythematosus (SLE) (5%). The UV syndrome is characterized by progressive uveitis and depigmenting dermatitis that closely resembles the human disease of VKH syndrome [1].

## 2. Case presentation

A 25 year old male patient with a known VKH syndrome, developed an acute onset of explosive diffuse joint pain and swellings and bilateral buttock pain two weeks after an attack of Chlamydial urethritis. Patient's history revealed bilateral buttock pain, and axial pain extending from the dorsal down to the lumbar spine which is typically inflammatory in nature and associated with axial morning stiffness lasting for one hour. Moreover the patient presented with symmetric pain and swelling affecting the small joints of both hands, both wrists, both ankles, and both knees.

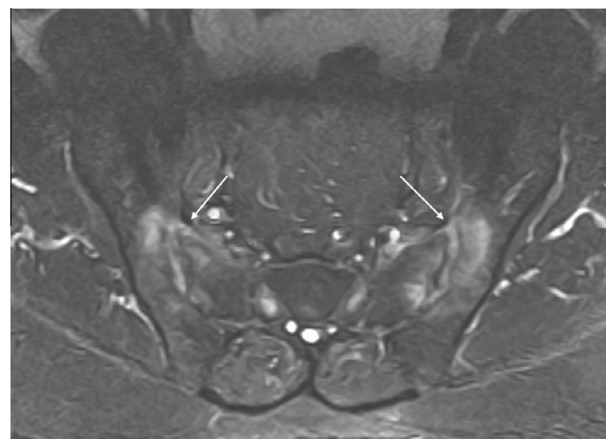
Rheumatological examination revealed symmetric form of arthritis involving proximal interphalangeal (PIP) joints, metacarpophalangeal (MCP) joints, both wrists, both knees and both ankles. Bilateral sacroiliitis was evident by direct sacral compression test and wide spread enthesopathy was also detected by careful clinical examination in the following areas; tender spinous processes extending from the dorsal to lumbar spine, bilateral anterior superior iliac spine, planter surface of both feet at the origin of plantar fascia and at the insertion of both tendo Achilles of both heels. The patient was treated two years before this event from autoimmune uveitis and diagnosed at that time as a case of VKH syndrome.

Additionally clinical examination revealed typical features of the disease with bilateral panuveitis, alopecia, poliosis, vitiligo and sensorineural hearing loss that was documented by audiometry examination tests. Interestingly the family history revealed that the mother of the patient had systemic sclerosis (diffuse type). His initial laboratory investigation revealed elevated ESR 1st h 60 mm/h, CRP 11 mg/DL, and negative rheumatoid factor (RF).

MRI of both sacroiliac joints showed evidence of bilateral sacroiliitis with extensive bone marrow edema of sacroiliac joints (Fig. 1). The patient was diagnosed as a case of reactive arthritis after an attack of chlamydial urethritis. A full course of Azithromycin to treat the underlying Chlamydial urethritis was received together with systemic steroids in a dose of 30 mg/day and methotrexate in a dose of 20 mg intramuscular on a weekly basis to control the reactive arthritis and the patient showed clinical improvement.

## 3. Discussion

VKH syndrome is an uncommon disorder characterized by uveitis, neurologic and dermatologic abnormalities, including tinnitus, vertigo, headache, meningoencephalitis, vitiligo, alopecia, and poliosis. The VKH syndrome has been reported to occur in association with other autoimmune disorders [1].



**Figure 1** Axial STIR showing bilateral sacroiliac joint effusion with subchondral bone marrow edema (white arrows).

The occurrence of such explosive form of reactive arthritis following an attack of Chlamydial urethritis in a case of established VKH syndrome is interesting and we do believe that our case was the first to report such rare association. Additionally the positive family history of scleroderma would explain the abnormalities of immunologic milieu that can take place in autoimmune disorders.

The uveal tract, although comprising three anatomic sections; the iris, the ciliary body and the choroid, may be regarded as a single functional unit, that is easily affected by several pathogenic agents that may act on infectious, toxic and immune basis. Since such inflammatory stimuli do not usually locate primarily in the eye but rather elsewhere in the body, uveitis is often associated with systemic diseases. This would explain the diversity of autoimmune disorders frequently associated with anterior or posterior uveal inflammation, such as Behçet's disease, Vogt-Koyanagi-Harada's syndrome, Reiter's syndrome, juvenile idiopathic arthritis, ankylosing spondylitis, sarcoidosis, Sjogren's syndrome, ulcerative colitis, psoriasis, tuberculosis and syphilis [2].

In other reports VKH syndrome was associated with other forms of seronegative spondyloarthropathy and most cases described an association of VKH syndrome with ulcerative colitis. Iversen and Sverrisson [3] were the first to report VKH syndrome in association with ulcerative colitis in a 32 year-Old Norwegian woman. More recently Federman et al. [4] described a case with established severe ulcerative colitis that developed VKH syndrome following traumatic brain insult. Most interesting, is that the authors reported the occurrence of severe reactive arthritis in the same case. Nevertheless de la Poza Gómez et al. [5] described a female patient who was diagnosed with VKH disease at the age of 14 years and consequently developed myelopathy, resulting in paraparesis, with associated demyelinating brain lesions in the periventricular white matter. Twelve years later, ulcerative colitis was diagnosed during workup for abdominal pain associated with bloody diarrhea in the same case.

This previous association of VKH syndrome with ulcerative colitis in previous reports [3–5] and reactive arthritis in our case that followed Chlamydial urethritis is interesting and deserves precise discussion and interpretation. Importantly in a previous report Federman et al. [4] described the occurrence

of severe form of reactive arthritis in their case together with ulcerative colitis. Axial affection in the form of bilateral sacroiliitis as described in our case further raises the possibility that VKH syndrome may be indeed a form of seronegative spondyloarthropathy (SpA).

Moreover recent data from HLA typing showed that HLA-DRB1 alleles encoding the shared epitope may act as an additional susceptibility factor for the development of SpA in HLA-B27-positive Japanese individuals [6]. Interestingly Patients with VKH had significantly a greater incidence of HLA-DRB1 \*0405 when compared to age and sex-matched controls, given that this finding suggests that HLA-DRB1 \*0405 allele might play a role in the pathogenesis of VKH disease [7].

Taken together the same epitopes that genetically predispose to VKH syndrome are the same that predispose to SpA groups of disorders and this explains the occurrence of reactive arthritis in our case and the previous reports that described the association between VKH syndrome and the occurrence of ulcerative colitis [3–5].

Although our concept is theoretical one, yet examining more cases with VKH syndrome looking for sacroiliac joint involvement by sensitive modalities like MRI in view of their HLA genetic back grounds may make our theory close to reality.

As of this writing, to our knowledge, this is the first case report in the literature that described an association between VKH syndrome and post Chlamydial urethritis reactive arthritis.

## Conflict of interest statement

All the authors declare no conflict of interest.

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